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SPONSOR TESTIMONY
SENATE BILL 173
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Chairman Burke, Vice Chair Huffman, Ranking Member Antonio and members of the Senate Health, Human Services & Medicaid Committee. Thank you for the opportunity to present to you sponsor testimony on Senate Bill 173, legislation that seeks to Designate September as Hirschsprung's Disease Awareness Month.

Between the 4th and the 12th weeks of pregnancy, while the fetus is growing and developing, nerve cells form in the digestive tract, beginning in the mouth and finishing in the anus. For unknown reasons, the nerve cells do not grow past a certain point in the intestine in babies with Hirschsprung's (HIRSH-sproongz) disease.¹

Hirschsprung's disease is a condition that affects the large intestine (colon) and causes problems with passing stool. The condition is present at birth as a result of missing nerve cells in the muscles of the baby's colon. In an infant with Hirschsprung's, stool moves through the bowel until it reaches the part where the nerve cells are gone. At that point, the stool moves slowly or stops.²

Infants with Hirschsprung's disease will very often have abdominal swelling (distention), abdominal pain, and vomiting. Affected infants have constipation and often exhibit poor weight gain, and slow growth.

Hirschsprung disease that occurs as an isolated problem has been associated with mutations in several different genes. Approximately 50% of affected individuals have one of these gene abnormalities. Hirschsprung disease affects males 3 to 4 times more often than females, although long-segment HSCR has a gender ratio of 1:1. The disorder occurs in approximately one in 5,000 live births. It is usually apparent shortly after birth, but may present in older children and adults.³

¹ <https://www.stanfordchildrens.org/en/topic/default?id=hirschsprungs-disease-90-P01999>

² <https://www.reachhd.org/#intro-1>

³ <https://rarediseases.org/rare-diseases/hirschsprungs-disease/>

Signs and symptoms of Hirschsprung's disease vary with the severity of the condition. Usually signs and symptoms appear shortly after birth, but sometimes they're not apparent until later in life. Typically, the most obvious sign is a newborn's failure to have a bowel movement within 48 hours after birth. Other signs and symptoms in newborns may include: Swollen belly; Vomiting, including vomiting a green or brown substance; Constipation or gas, which might make a newborn fussy and Diarrhea.⁴

Children who have Hirschsprung's disease are prone to a serious intestinal infection called enterocolitis. Enterocolitis can be life-threatening and requires immediate treatment.

In almost all cases, treatment of Hirschsprung disease requires surgery to remove the part of the colon and/or rectum that lacks the normal nerve development, and to join the two healthy ends together. There are three standard surgical procedures designed to correct this disorder. Each procedure removes the affected part and attaches the healthy part of the bowel to the rectum completing what is known as a "pull-through" procedure. Currently most procedures are performed in a single stage.

What this measure seeks to do is increase public awareness of Hirschsprung's disease and encourage caregivers of newborns to seek an early diagnosis and proper treatment of the disease.

Chairman Burke and members of the committee. Thank you for the opportunity to present to you sponsor testimony on Senate Bill 173. I am happy to answer any questions that you might have.

⁴ <https://www.mayoclinic.org/diseases-conditions/hirschsprungs-disease/symptoms-causes/syc-20351556>